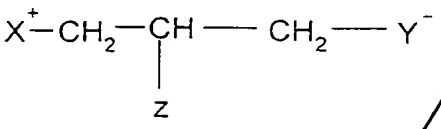
CLAIMS

1) Compounds of formula (I)



wherein: X^+ is selected from the group consisting of $N^+(R_1,R_2,R_3)$ and $P^+(R_1,R_2,R_3)$,

wherein (R_1,R_2,R_3), being the same or different, are selected in the group consisting of hydrogen and C_1-C_9 straight or branched alkyl groups, -CH=NH(NH_2), - NH_2, -OH; or two or more R_1, R_2 and R_3, together with the nitrogen atom, which they are linked to, form a saturated or unsaturated, monocyclic or bicyclic heterocyclic system; with the proviso that at least one of the R_1, R_2 and R_3 is different from hydrogen;

Z is selected from

-OR_4,

-$OCOOR_4$,

-$OCONHR_4$,

-$OCSNHR_4$,

-$OCSOR_4$,

-NHR_4,

-$NHCOR_4$,

-$NHCSR_4$,

-$NHCOOR_4$,

-$NHCSOR_4$,

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-$NHCONHR_4$,

-$NHCSNHR_4$,

-$NHSOR_4$,

-$NHSONHR_4$,

-$NHSO_2R_4$,

-$NHSO_2NHR_4$,

-SR_4,

wherein -R_4 is a C_1-C_{20} saturated or unsaturated, straight or branched alkyl group, optionally substituted with a A_1 group, wherein A_1 is selected from the group consisting of halogen atom, aryl, heteroaryl, aryloxy or heteroaryloxy group, said aryl, heteroaryl, aryloxy or heteroaryloxy groups being optionally substituted with one or more C_1-C_{20} saturated or unsaturated, straight or branched alkyl or alkoxy group and/or halogen atom;

Y^- is selected from the group consisting of $-COO^-$, PO_3H^-, $-OPO_3H^-$, tetrazolate-5-yl;

with the proviso that when Z is -$NHCOR_4$, X^+ is trimethylammonium, Y^- is $-COO^-$, then R_4 is C_{20} alkyl;

with the proviso that when Z is -$NHSO_2R_4$, X^+ is trimethylammonium and Y^- is $-COO^-$, then R_4 is not tolyl;

with the proviso that when Z is -NHR_4, X^+ is trimethylammonium and Y^- is $-COO^-$, then R_4 is not C_1-C_6 alkyl.

their (R,S) racemic mixtures, their single R or S enantiomers, their pharmaceutically acceptable salts .

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2) Compounds according to claim 1, wherein R_1, R_2 and R_3 are methyl.

3) Compounds according to claim 1, wherein the heterocyclic system formed by R_1, R_2 and R_3 together with nitrogen is selected from the group consisting of morpholinium, quinuclidinium, pyridinium, quinolinium and pyrrolidinium.

4) Compounds according to claim 1, wherein R_1 and R_2 are H, R_3 is selected from the group consisting of -CH=NH(NH_2), - NH_2 and – OH.

5) Compounds according to ~~any one of claims 1-4,~~ claim 1 wherein Z is selected from the group consisting of ureido (-NHCONHR_4) or carbamate (-OCONHR_4, -NHCOOR_4), R_4 is a C_7-C_{20} saturated or unsaturated, straight or branched alkyl group.

6) Compounds according to claim 5, wherein R_4 is a C_9-C_{18} saturated or unsaturated, straight or branched alkyl group.

7) Compounds according to claim 1, selected from the group consisting of

R,S-4-trimethylammonium-3-(nonylcarbamoyl)-aminobutyrate;

R,S-4-quinuclidinium-3-(tetradecyloxycarbonyl)-oxybutyrate;

R,S-4-trimethylammonium-3-(nonylcarbamoyl)-oxybutyrate;

R,S-4-trimethylammonium-3-(nonyloxycarbonyl)-oxybutyric acid chloride;

R,S-4-trimethylphosphonium-3-(nonylcarbamoyl)-oxybutyrate;

R,S-4-trimethylammonium-3-(octyloxycarbonyl)-aminobutyrate;

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R,S-4-trimethylammonium-3-(nonyloxycarbonyl)-aminobutyrate;

R,S-4-trimethylammonium-3-octyloxybutyrate;

R,S-4-trimethylammonium-3-tetradecyloxybutyrate;

R,S-1-guanidinium-2-tetradecyloxy-3-(tetrazolate-5-yl)-propane;

R,S-1-trimethylammonium-2-tetradecyloxy-3-(tetrazolate-5-yl)-propane;

R,S-3-quinuclidium-2-(tetradecyloxycarbonyl)-oxy-1-propanephosphonate monobasic;

R,S-3-trimethylammonium-2-(nonylaminocarbonyl)-oxy-1-propanephosphonate monobasic;

R,S-3-pyridinium-2-(nonylaminocarbonyl)-oxy-1-propanephosphonic acid chloride;

R-4-trimethylammonium-3-(tetradecylcarbamoyl)-aminobutyrate;

R-4-trimethylammonium-3-(undecylcarbamoyl)-aminobutyrate;

R-4-trimethylammonium-3-(heptylcarbamoyl)-aminobutyrate;

R,S-4-trimethylammonium-3-(nonylthiocarbamoyl)-aminobutyrate;

R-4-trimethylammonium-3-(nonylcarbamoyl)-aminobutyrate;

S-4-trimethylammonium-3-(nonylcarbamoyl)-aminobutyrate;

S-4-trimethylammonium-3-(tetradecylcarbamoyl)-aminobutyrate;

R,S-4-trimethylammonium-3-tetradecylaminobutyrate;

R,S-4-trimethylammonium-3-octylaminobutyrate;

R,S-4-trimethylammonium-3-(decansulfonyl)aminobutyrate;

R,S-4-trimethylammonium-3-(nonylsulfamoyl)aminobutyrate;

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S-4-trimethylammonium-3-(dodecansulfonyl)aminobutyrate;

R-4-trimethylammonium-3-(dodecansulfonyl)aminobutyrate;

S-4-trimethylammonium-3-(undecylsulfamoyl)aminobutyrate;

R-4-trimethylammonium-3-(undecylsulfamoyl)aminobutyrate;

R-4-trimethylammonium-3-(dodecylcarbamoyl)aminobutyrate;

R-4-trimethylammonium-3-(10-phenoxydecylcarbamoyl)aminobutyrate;

R-4-trimethylammonium-3-(*trans*-β-styrenesulfonyl)aminobutyrate.

8) A process for the preparation of compounds of claim 1, wherein Z is carbonate (-OCOOR$_4$), carbamate (-NHCOOR$_4$, -OCONHR$_4$), thiocarbamate (-OCSNHR$_4$, -NHCSOR$_4$) or thiocarbonate (-OCSOR$_4$), comprising the reaction of X+-CH$_2$-CH(OH)-CH$_2$-Y-, wherein X+ and Y- have the same meanings as in claim 1, of the desired structure, optionally protected on the acid Y- group, respectively with alkyl chloroformates, alkyl isocyanates, alkyl isothiocyanates, alkyl thiochloroformates, wherein the alkyl moiety is the desired R$_4$ alkyl group.

9) A process for the preparation of the compounds of claim 1, wherein Z is amide (-NHCOR$_4$), thioamide (-NHCSR$_4$), carbamate (-NHCOOR$_4$, -OCONHR$_4$), thiocarbamate (-NHCSOR$_4$, -OCSNHR$_4$), ureido (-NHCONHR$_4$), thioureido (-NHCSNHR$_4$), sulfinamide (-NHSOR$_4$), sulfonamide (-NHSO$_2$R$_4$), sulfinamoylamino (-NHSONHR$_4$), and sulfamide (-NHSO$_2$NHR$_4$), comprising the

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reaction of X^+-CH_2-$CH(NH_2)$-CH_2-Y^-, wherein X^+ and Y^- have the same meanings as in claim 1, of the desired structure, optionally protected on the acid Y^- group, respectively with acyl chlorides, thioacyl chlorides, alkyl chloroformates, alkyl thiochloroformates, alkyl isocyanates, alkyl thioisocyanates, alkyl sulfinyl chlorides, alkyl sulfonyl chlorides, $SOCl_2$ and alkyl amines, alkyl sulfamoyl chlorides (or SO_2Cl_2 and alkyl amines), wherein the alkyl moiety is the desired R_4 alkyl group.

10) A process for the preparation of the compounds of claim 1, wherein Z is –OR_4 or -SR_4 comprising

a) the reaction of carbonyl compounds of formula Hal-CH_2-CO-CH_2-COOR' , wherein Hal is a halogen atom and R' is the residue of a suitable ester, with respectively alcohols and thiols R_4OH or R_4SH, wherein R_4 is as defined in claim 1, to give the respective ketal or thioketal;

b) transformation of the the respective ketal or thioketal into the respective ether or thioether;

c) substitution of the Hal atom with a nucleophilic group, and

d) transformation of the nucleophilic group into the X+ group, wherein X^+ is $N^+(R_1,R_2,R_3)$ or, alternatively

e) step b) is followed by the substitution of the Hal atom with a (R_1,R_2,R_3)-substituted phosphine to obtain the compounds of formula (I) wherein X^+ is $P^+(R_1,R_2,R_3)$.

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11) A process for the preparation of the compounds of claim 1, wherein Z is $-NHR_4$ comprising the reaction of $X^+-CH_2-CH(NH_2)-CH_2-Y^-$, wherein X^+ and Y^- have the same meanings as in claim 1, of the desired structure, optionally protected on the acid Y^- group, with alkane carbaldheydes, wherein the alkyl moiety is a one-term lower homologue of the desired R_4, and subsequent reduction.

12) Compounds according to ~~claims 1-7,~~ claim 1 for use as medicaments.

13) Pharmaceutical composition comprising a therapeutically effective amount of at least a compound of ~~claims 1-7~~ claim 1, in admixture with pharmaceutically acceptable vehicles and excipients.

14) Pharmaceutical composition comprising a therapeutically effective amount of at least a compound of ~~claims 1-7~~ claim 1, in admixture with pharmaceutically acceptable vehicles and excipients and optionally in combination with other active ingredients.

15) Use of a compound of ~~claims 1-7~~ claim 1, for the preparation of a medicament useful for the treatment of pathologies related to a hyperactivity of carnitine palmitoyl-transferase.

16) Use according to claim 15, wherein said pathology is selected from the group consisting of hyperglycaemia, diabetes and pathologies related thereto, heart failure, ischemia and ketonic states.

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17) Pharmaceutical composition according to claim 14, wherein said other active ingredient is a suitable well-known active ingredient for the treatment of diabetes.

18) Pharmaceutical composition according to claim 17, wherein said other active ingredient suitable for the treatment of diabetes is selected from the group consisting of sulfonylurea, L-carnitine, fibrate and other agonists of peroxisomal proliferator activated receptor (PPAR-α), agonists of 9-cis retinoic acid activated receptor, HMG-CoA reductase inhibitor, β-sitosterol inhibitor, cholesterol acyltransferase inhibitor, biguanides, cholestyramine, angiotensin II antagonist, melinamide, nicotinic acid, fibrinogen receptor antagonists, aspirin, α-glucosidase inhibitors, insulin secretogogue, insulin and glucagon-like peptides (incretins) and agonists of PPAR-γ.

19) Use of the pharmaceutical composition ~~any one of claims 17-18~~ claim 17 for the treatment of diabetes.

20) Pharmaceutical composition according to claim 14, wherein said other active ingredient is a suitable well-known active ingredient for the treatment of obesity.

21) Pharmaceutical composition according to claim 20, wherein said other active ingredient suitable for the treatment of obesity is selected from the group consisting of fenfluramine, dexfenfluramine, phentiramine, a β-3-adrenergic receptor agonist.

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22) Use of the pharmaceutical composition ~~any one of claims 20-21~~ claim 20 for the treatment of obesity.

23) Pharmaceutical composition according to claim 14, wherein said other active ingredient is a suitable well-known active ingredient for the treatment of high triglyceridhemia.

24) Pharmaceutical composition according to claim 14, wherein said other active ingredient suitable for the treatment of high cholesterol levels and in modulating HDL plasma levels.

25) Pharmaceutical composition according to claim 24, wherein said active ingredient suitable for the treatment of high cholesterol levels and in modulating HDL plasma levels, is selected from the group consisting of fibrates, and other PPAR-α agonists; inhibitors of cholesterol biosynthesis, HMG-CoA reductase inhibitors, statins, inhibitors of cholesterol absorption, acyl CoA:cholesterol acyltransferase inhibitors, anion exchange resins, nicotinyl alcohol, nicotinic acid or a salt thereof; vitamin E; thyromimetics and L-carnitine.

26) Use of the pharmaceutical composition ~~any one of claims 24-25~~ claim 24 for the treatment of high cholesterol levels and related diseases.

27) Use according to claim 26 for the treatment of hypertension, obesity, atherosclerosis, diabetes and related conditions.

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